

Surgical Repair of a Median Cleft of the Upper Lip via a Pfeifer Incision: A Case Report

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Abstract

Median cleft is the midline cleft of the lip. It develops due to incomplete or failed fusion of the median nasal prominence. It can present with minimal deformities such as involvement of the vermilion border, or complex clefting of the midline structures and brain. Median clefts are broadly classified as true and false clefts. This case report describes a rare case of median cleft of the upper lip involving the white roll, which was not associated with any other deformities. Treatment included reconstruction of the philtrum and the cupid's bow while maintaining vermilion fullness and continuity, and minimizing scar formation. Various techniques have been advocated for treatment of this type of median upper lip cleft. Here we describe a technique using Pfeifer incision to correct our patient's defect. Pfeifer incision consists of wavy lines and its use has been advocated for correction of various craniofacial abnormalities.

Keywords: Median Cleft Lip; Hypertelorism; Craniofacial Abnormalities

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INTRODUCTION

Craniofacial defects are rare, disfiguring facial anomalies, with an incidence of about 1.4 to 4.9 cases per 100,000 live births [1]. Craniofacial clefts may be caused by failure of the maxillary processes to fuse, external pressure, amniotic bands, oligohydramnios, central disorganization of the neural crest, and hematomas [2]. In 1973, Tessier [3] classified craniofacial clefts based on clinical, radiographic, and surgical findings. The median cleft of the upper lip, or Tessier 0 class, has a variety of presentations, ranging from a minimal notching of the lip, vermilion, and nose to a wide cleft that divides craniofacial structures [4]. Tessier 0 clefts result from failure of the two medial nasal processes to fuse at midline [5]. The incidence of Tessier 0 clefts is reported to be about 0.43% to 0.73% [6]. Median clefts are broadly classified into true and false; false clefts are due to agenesis of the medial nasal process, while true clefts are due to failure of the medial nasal process to fuse. Treatment of median clefts

depends on the clinical presentation and may vary from simple alignment of orbicularis oris and vermilion, to reconstruction of the cupid's bow and philtrum for true median clefts, and complex craniofacial procedures in case of false median clefts. Various techniques have been described to repair mild or moderate true median cleft lips. Here we report using a Pfeifer incision to correct a moderate true median cleft of the upper lip. Pfeifer incision includes wavy lines, which elongate the incised tissues as the waves are straightened to close in a straight line, and also provide extra tissue for a tension free closure.

CASE REPORT

A five-year-old boy with a facial cleft presented to our clinic. On examination, there was a median cleft of the upper lip involving the white roll, with no bony involvements. The child was diagnosed with Tessier type 0 craniofacial cleft (Fig. 1).



Fig. 1: Median cleft of the upper lip



Fig. 2: Incision marking for Pfeifer technique (A,A': Highest points on white roll; B,B': Deepest points on white roll)

The cleft was repaired using a Pfeifer incision. The highest- (points A, A'), and the deepest-points (points B, B') of the white roll were marked on both sides. Subsequently, a wavy incision line was made starting from the deepest point and extending over the philtrum just above the cleft (Fig. 2). A diamond incision was made over the vermillion and the labial mucosa, intra-orally extending just beyond the cleft margin. Incision was made and the mucosal tissue covering the area medial to the incision site (sterile zone) was removed. Undermining of the orbicularis oris muscle was performed and it was approximated using 4-0 vicryl. Skin was closed in a straight line using 6-0 ethilon sutures. Postoperatively, the patient had a satisfactory result. The cupid's bow was properly aligned, with equal height on both sides (Fig. 3). Vermilion form was satisfactory and the fullness and continuity of the orbicularis oris were maintained.

DISCUSSION

Tessier 0 clefts are the most common form of craniofacial clefts [7]. A median cleft can also be called midline cleft or vertical cleft through the centre of the upper lip, and is a rare anomaly, the exact developmental origin of which is not clear. Tessier 0 clefts occur during the third week of gestation due to failure of the two medial nasal processes to close in midline. It can occur in isolation or be a part of a syndrome such as orofacial-digital syndrome.

Most patients with severe false clefts do not survive. False clefts are associated with abnormalities of the forebrain and are categorized under the category of holoprosencephaly [8,9]. In 1937, Veau [10] categorized median clefts to notch of the lip, median cleft extending to the columella, and defects due to atrophy of the midline facial structures. Median cleft face syndrome, fronto-nasal dysplasia, and Tessier 0 clefts are various terms used to describe abnormalities associated with true median clefts that are not accompanied by forebrain abnormalities [3]. The tessier 0 anomaly may present as a small notch in the soft tissue, or in association with hypertelorism, midline craniofacial osseous defects, and hairline abnormalities. The majority of cases are sporadic, but familial cases have also been documented [11]. Severe cases require reconstruction of the nasal anatomy. Correction of hypertelorism is best delayed until the patient is eight years old.



Fig. 3: Postoperative result after suture removal

Various treatment options are present for mild deformities which do not involve the white roll. When developing the treatment plan, reconstruction of the cupid's bow, labial philtrum, vermillion, and buccal mucosa should be kept in mind. Urata and Kawamoto [12] described using a V-Y flap, while Weimer et al, [13] used a diamond incision to repair these anomalies. Da Silva Frietas and colleagues [14] described a mucosal Z transposition technique to treat mild cases. For moderate defects involving the philtrum, Millard and Williams [15] reported using an inverted V excision. In this case report we used a Pfeifer incision to reconstruct a moderate true midline cleft.

Pfeifer incision consists of short curved waves, which are made around the defect. These waves are subsequently approximated in a straight line, which help expanding the length and width of the tissue. This incision has previously been used to correct other Tessier clefts with a high success rate. It has also been used for palatal repair with promising results. The above-mentioned studies prove the versatility of the Pfeifer incision. In our case we also found that the Pfeifer incision is very easy to execute. The incision helped in tension free closure of the cleft tissues. It also helped to properly align the orbicularis oris and the white roll. We were able to achieve adequate symmetry of the philtrum and the cupid's bow, which is important in cases like this.

The postoperative results were excellent and proper approximation of the orbicularis oris muscle and the vermillion was achieved with a symmetrical cupid's bow and philtrum. The patient's scar healed well with no evidence of hypertrophy. The only disadvantage of this technique is that the final closure line is placed directly over the philtrum.

CONCLUSION

Since the presentation of midline cleft deformities varies widely, each case should be individually considered and treated. Pfeifer incisions can successfully provide mucosal

length, vermillion fullness, and lip symmetry in patients with moderate median cleft lip.

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